WHAT IS MENIERE'S DISEASE?

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"Our hearing is a choice and dainty sense and hard to mend yet soon it may be marred. These are the things that breedeth most offence". This quotation from the Regimen Sanitatus Salernitatum illustrates that the ancient physicians knew what outside influences harmed the organ of hearing and the effect on day-to-day living. However, the anatomy of the ear remained a mystery until the work of Antonio Scarpa. In 1789 Scarpa published the results of his dissections, accompanied by beautiful engravings, and there can be few more eloquent reasons for naming the spiral ganglion after him. Scarpa was professor of anatomy and surgery in Modena and Paris and in Vienna. At the same time Dominico Cartugno discovered that the inner ear was not filled with air as had been previously thought, but contained fluid. It was Scarpa who pointed out the difference between perilymph and endolymph and separated the structures of the saccule and the utricle.

In England at about the same time, the work of John Cunningham Saunders heralded the dawn of modern otology. In 1806 he published his treatise on diseases of the ear, illustrating it with dissections. He denoted sensorineural deafness as "a want of sensibility in the nerve, some alteration in the structures of the membranes on which the nerve is expanded, or change in the properties of that fluid which is contained in the membranes and is the immediate agent in impressing the sensitive extremities of the nerve".

Sir William Wilde, better known as the father of Oscar, was influenced by Saunders. In his thesis on the medical history of Jonathan Swift, the author of Gulliver's Travels, he documents the various stages of what is now patently Swift's Meniere's disease. He describes an increasing fluctuating deafness, tinnitus and an associated vertigo. Swift's physicians at the time thought the underlying cause was united but could not accurately localise the problem[1]. Shakespeare mentions the association of deafness and "falling sickness" in Julius Caesar, and there is no doubt that the syndrome

was previously recognised although attributed to a cerebral rather than a labyrinthine disorder. Meniere's great contribution to the disease that bears his name was proving that the symptoms are of labyrinthine origin and not due to brain disease as formerly thought. In his series of 5 articles in the Medical Journal of Paris in 1861, Meniere[2] mentioned the characteristic repeated attacks of vertigo with nausea and vomiting occurring during weeks, months or years in an otherwise healthy person except for hearing impairment, sometimes bilateral often unilateral. Meniere also noted that the hearing loss was of the neurological type but involved the low frequencies more than the high and was accompanied by tinnitus. He related the case report of a young girl who, while riding on top of a stagecoach, suffered a sudden onset of deafness, vertigo and vomiting. She died 5 days later and at post-mortem it was found she had had an apparent haemorrhage into the labyrinth with no central nervous system abnormality[3, 4]. The aetiology and pathology of Meniere's disease remained obscure for many years. In 1871 Knapp[5], an ophthalmologist-otologist compared Meniere's disease to glaucoma of the eye, suggesting the symptoms may be due to an increase in the pressure in the labyrinth.

ANATOMY

The inner ear consists of 3 main parts; the membranous (otic) labyrinth which is filled with endolymph; the osseous (periotitic) labyrinth and the otic capsule surrounding the osseous labyrinth.

The membranous labyrinth is an endolymph containing system of epithelial tubes and spaces. The cochlear duct follows the spiral of the bony cochlea from the vestibular caecum in the cochlear recess of the vestibule to the cupular caecum, which helps form the helicotrema. Adjacent to the cochlear duct is the saccule. It is located in the spherical recess in the anterior-inferior part of the vestibule and is connected to the cochlear duct by the

ductus reuniens. From the saccule runs the endolymphatic duct which passes through the bony aqueduct of the vestibule to end as a dilated saccus endolymphaticus external to the dura mater. The saccus endolymphaticus is a relatively large organ, usually as long as the distance from the posterior semicircular canal to the anterior edge of the cochlea. From the surgical point of view, following a mastoidectomy, the sac may be found between the sigmoid sinus and the posterior semicircular canal at or below a line passing through the horizontal semicircular canal. Great variations in landmarks must, however, be expected. Likewise the sac itself may be very variable - either thin walled and full of fluid, or the walls may be contiguous or even adherent. The size of the sac is very variable such that on occasion even the most experienced of operators may have difficulty in accurate localisation of the sac.

OSSEOUS LABYRINTH

The osseous labyrinth is the space occupied by the periotitic labyrinth and is filled with perilymph. Contrary to what its name suggests it is not a bony structure.

The cochlea is conical; its base is located on the internal acoustic meatus and is about 9 mm across. The cupula is directed anterolaterally with a height of about 5 mm. It forms a spiral tube about 30 mm in length passing through $2\frac{3}{4}$ turns around the modiolus. The osseous spiral lamina projects from the modiolus, separating the scala of the vestibule laterally from the scala tympani medially. The modiolus, of course, contains longitudinal and spiral canals conducting nerves and vessels.

The scala vestibuli is an extension of the vestibule accompanying the cochlear duct from which it is separated by Reissner's membrane throughout its course. At the helicotrema the scala vestibuli communicates with the scala tympani from which it is separated throughout its parallel course by the osseous spiral lamina. and the basilar membrane

OTIC CAPSULE

Surrounding the osseous (periotitic) labyrinth is the otic capsule. It is a 3 layered structure of bone and has a number of communications. The vestibular window allows communication with the vestibule, and the cochlea window with the scala tympani. The vestibular aqueduct transmits the endolymphatic duct from the internal opening in the elliptical recess to the external opening. The cochlear canaliculus is usually a potential space although it can be patent. Its internal window is near the cochlear window and it passes from there to an external opening medial to the jugular fossa. The fissula ante fenestram is an appendage of the periotitic labyrinth extending from the vestibule anterior to the vestibular window to the periostium of the tympanic cavity.

It must be remembered, however, that the hallmark of the temporal bone is variation[6].

PATHOPHYSIOLOGY

In 1938, Hallpike and Cairns published the histopathological findings in temporal bones of 2 patients with Meniere's disease who had both died soon after posterior fossa VIII nerve section. This was the first description, in English, of the idiopathic hydrops of the pars inferior, characteristic of the disease[7]. Mygind and Dederding, however, again compared the disease with glaucoma. They suggested the lesion was a hydrops of the labyrinth produced by capillary dysfunction with a resultant defective oxidation and increase in water-binding efficiency in the cells and with subsequent intracellular oedema[8]. These findings were later confirmed by many other workers and the term ENDOLYMPHATIC HYDROPS came to be used synonymously with Meniere's disease[9, 10, 11, 12, 13]. The principle finding by Friedmann[14] was that of distension of the cochleo-saccular endolymphatic system at post-mortem in the

labyrinth of those patients who had suffered from Meniere's disease during life. The utricle was found to be less dilated and the semicircular canals normal[15, 16]. Hydrops of the membranous inner ear can, however, occur in late syphilis[17] and in diseases of the otic capsule, such as otosclerosis and Paget's disease of bone[18-23]. Post inflammatory hydrops has been described with bony obliteration of the labyrinth[24]. As the knowledge of the pathology of the temporal bone increases, no doubt other aetiological factors will come to light. The idiopathic group is, therefore, what is termed Meniere's disease.

The pathogenesis of Meniere's disease has not been definately established. It is accepted that endolymphatic hydrops is the most significant change in the inner ear, but the problem of the causation of the hydrops remains undetermined. The major change, it is agreed, is the gross dilatation of the scala media and saccule. Reissner's membrane bulges into the scala vestibule and may eventually rupture, allowing perilymph to mingle with endolymph. After rupture the pressures are equalised and the membrane may proceed to repair itself[25, 31]. These ruptures may occur at any site in Reissner's membrane or the saccule, utricle and ampulla[16]. Rupture at the membranous labyrinth would explain most reasonably the episodic and paroxysmal nature of Meniere's disease. The saccular wall and Reissner's membrane appear to be more yielding than the walls of the utricle and canals and, therefore, show more distension and fewer ruptures. Thus the saccular wall and Reissner's membrane often come to lie against the bony walls. A total collapse of the membranous labyrinth in Meniere's disease may occur as a result of failure of a fistula to heal thus not allowing endolymph to reaccumulate. Another interesting histological feature that is occasionally observed in Meniere's disease is degeneration of the sensorineural elements in the apical region of the cochlea. The occurrence of fibrosis around the vestibule was first noted by Hallpike and Cairns in 1938 and has been confirmed many times since[16, 25, 30]. In cases of Meniere's disease it has been suggested that the endolymphatic sac is ischaemic and the lumen is often reduced or obliterated by adhesions of the walls, while the sac is often

small and sometimes cannot be found at the usual location directly behind the horizontal semicircular canal[26]. The endolymphatic duct may be clearly seen by polytomography in most normal ears, but in Meniere's disease the duct is, more often than not, filiform in appearance or not seen at all[27-29]. A giant among men, Georges Portmann in 1927, spoke of Meniere's disease as an "aural glaucoma"[32] and for the first time suggested that deficient resorption of endolymph by the endolymphatic sac might be the cause. Retzius among other anatomists had mentioned the endolymphatic sac without attaching any importance to it, but Portmann[33], working successively on elasmobranchs, teleosts, frogs, birds and mammals, found the organ constant. He then blocked off the communicating channel between the sac and the sea-water in the Ragides, which produced vertigo in the fish in various swimming positions. Kimura and Schuknecht[34] consistently repeated these observations in guinea pigs by obstructing the endolymphatic duct. Beal[87] obtained similar results in rabbits and cats. Kimura[35] subsequently demonstrated that the scala media volume increased by, on average, 40% during the first 24 hours after ablation of the duct and thereafter slowly enlarged over several months. Eventually the saccule became grossly distended and atrophic changes took place in the stria vascularis and neurosensory structures of the cochlear apex. The pars superior remained relatively normal.

It has since been ascertained that in different experimental animals, eg cats, rabbits and monkeys, much longer survival times are necessary before significant hydrops of the pars inferior develops. There is no concrete evidence as to what time interval is required in man, but the histopathological changes in human Meniere's disease are essentially similar to those in animal models[33,36].

In 1952, Von Bekesy[37] described the large positive DC potential in endolymph and in 1954, Smith et al[38] described the unusual high potassium and low sodium electrolyte composition of that fluid. In 1927, Guild[39] proposed that endolymph flowed from the stria vascularis, down through the ductus

reuniens, into the saccule and finally through the endolymphatic duct into the sac to be reabsorbed.

This proposal was based on studies of injection of substances into the endolymph and was subject to some criticism. It became known as the longitudinal flow theory. In 1960 it was reaffirmed by Ormerod[40] who demonstrated a heavy concentration of radio-active sulphur in the distal part of the endolymphatic duct in pigeons. On the other hand, Lindsay and others obliterated the endolymphatic duct and sac in monkeys and cats and found no build up of endolymph or any functional changes within the membranous labyrinth[41-43]. Another theory seemed to be needed. In 1958, Naftalin and Harrison[44] proposed the radial flow theory of endolymph reabsorption later supported by Lawrence and Wolsk[45]. This theory states that the endolymph is produced and reabsorbed in the same area - a radial type of flow from the stria vascularis to the hair cells and back to the stria vascularis. Lawrence reasoned that if longitudinal flow occurred, creating a hole in Reissner's membrane should allow the endolymph to leak out and thus damage the hearing below thepoint of rupture. They found that a hole, say at the 3 kH area, produced a drop in hearing at that point but no change from 3 - 10 kH. They, therefore, concluded that the radial flow theory was valid, but they also demonstrated that the endolymphatic and perilymphatic systems can be connected without serious consequences to the inner ear. The evidence for this conclusion came purely from the observation in animals that when Reissner's membrane breaks in an otherwise normal ear, perilymph enters the scala media. This observation was made in animals in which Reissner's membrane was broken either surgically or by loud sounds, not by a process of increased amounts of endolymph. In one series of experiments a special biologic stain was applied to the celloidin embedded section of a guinea pig ear that had been subjected to loud sounds. This ruptured the Reissner's membrane in every animal. The animals were kept in health for 2 months following the exposure at which time the temporal bones were processed.

Microscopic examination revealed mingling of perilymph with the darker stained endolymph only in the region of the membrane tear. The celloidin lost its darker stain within the scala media making it appear that perilymph had entered rather than endolymph leaving[31,46-48]. The interesting thing was that the stria vascularis and the organ of Corti degenerated in the area of this mixing. By electron microscopic studies Duvall[95] found marked atrophy of the stria vascularis and spiral ligament next to or slightly apical to the area of rupture. Earlier evidence had indicated that perilymph contamination of the scala media affects the organ of Corti both morphologically and functionally. Injection of artificial perilymph into the scala media by Davis et al[49] abolished the local cochlear microphonic. Bekesy reported that as he looked at the organ of Corti through Reissner's membrane in the living guinea pig, when perilymph flowed through a break in Reissner's membrane, the hair cells, which normally look like oil droplets, turned opaque[50]. Thus the evidence indicates that if perilymph enters the endolymphatic system, decreased function of the sensory area results. However, in animals even 2 months after rupture of Reissner's membrane, mingling of perilymph with endolymph occurred only in the region of the opening[45].

Ranch[94] has demonstrated the dynamic role of Reissner's membrane by using radio-active potassium and sodium. When K^{42} (KCl) is injected into perilymph it quickly appeared in the endolymph so that after about 2 minutes the concentration of K^{42} in the endolymph was greater. The potassium transport was 4-5 times faster than the sodium transport.

Wullstein and Ranch[51] demonstrated that the endolymph removed from a patient with Meniere's disease had a normally high potassium content. It will be recalled that endolymph has a high potassium content and perilymph a high sodium content. This, therefore, suggests the endolymph filling the system is normal fluid which in turn suggests inadequate reabsorption. For the basilar membrane to vibrate normally it must have the same fluid pressures on both sides. It would seem that if the inner ear is to function normally

it must have a scala vestibuli containing perilymph, a scala media with endolymph and an actively functioning Reissner's membrane between the 2 fluids. In decompensated Meniere's disease this system has broken down since Reissner's membrane is now distended to contact the wall of the scala vestibuli and it bulges out through the helicotrema. This situation may account for the continued deterioration in Meniere's disease. In 1957, Tonndorf demonstrated on a cochlear model that by mechanically increasing the endolymphatic pressure over the perilymphatic pressure, a low tone type of loss and diplacusis were produced, ie the same sound elicits a different pitch in the normal and affected ear, usually higher in the latter. Apparently since the basilar membrane is widest at the area of the low frequencies (apex), increased pressure distends the membrane more here and dampens its movement more than at the narrower high frequency portion. This pressure also shifts the area of the membrane that is vibrated by a given frequency and leads to a diplacusis type of distortion. McCabe and Wolsk[53] confirmed this observation in animals. They injected a high potassium, low sodium (endolymph-like) fluid directly into the scala media and demonstrated a decrease in the cochlear microphonics. Upon release of this pressure the microphonics returned to normal. The elasticity of the membranous labyrinth was confirmed by Henriksson[54] in frogs by injecting and recovering on release of pressure the same amount of fluid from the frog's endolymphatic system. Godlowski[104] believes that it is the local expansion of a generalised molecular disease of proteins affecting the whole organism and acting through a hyaluronidase deficiency within the bony labyrinth, to produce a mild hyperosmolarity and endolymphatic hydrops.

The electron microscope has ushered in a new wave of research material to unravel the complexities of Meniere's disease. In this context the first case was described by Pietrantoni and Iurata in 1960[55]. They noted in Meniere's disease the almost total loss of acoustic hairs of the sensory cells of the macula, associated with a spongy appearance of the much thinner cuticle and of the cytoplasm, due to the presence of cytoplasmic vacuoles. They considered,

of course, the possibility of tissue damage through poor presentation, but concluded that the absence of cilia, the cuticular changes and cytoplasmic vacuolation were genuine pathological features of the disease. In another case described by Litton and Lawrence[56], there was widespread degeneration of the epithelium. Ireland and Farkashidy[57] confirmed the previous findings and added a new feature in the shape of a banded structure, consisting of parallel bands of relatively opaque material and interpreted it as a degenerative change due to Meniere's disease. Friedmann, Cawthorne and Bird[58,59] discovered an unusual laminated structure in the degenerating neuroepithelium of patients operated on to cure Meniere's disease. This was confirmed by Hilding and House[60]. The overall impression is that of more or less widespread degeneration, both sensory and supporting cells showing vacuolation of the cytoplasm. Regenerative processes may alternate with the underlying degenerative process causing Meniere's disease. This concept helps to explain the remissions after an attack.

Notwithstanding all the preceding observations and experimental results, the aetiology and pathology of Meniere's disease remains obscure and no real progress is to be expected in this respect until we have a much more detailed knowledge of the vascular supply and innervation of the human labyrinth, the finer structure of the areas concerned with the production and reabsorption of the labyrinthine fluids, and the chemical composition of the latter under normal and pathological conditions[61].

CLINICAL FEATURES

Meniere's disease is one of the least understood disorders, by both specialists and doctors in general. It is variable in clinical presentation, imprecise in diagnosis, and because the effectiveness of treatment is doubtful, it leaves the clinician disillusioned, depriving the patient of the potential cure that some authors believe is possible[33]. It is important to

diagnose the condition of Meniere's disease accurately; to distinguish it from those other diseases which may masquerade under the same name. To attempt this clarification a list of positive and negative criteria has been suggested[105].

In 1972, the American Academy of Ophthalmology and Otolaryngology Committee on Equilibrium defined Meniere's disease as follows " a disease of the membranous inner ear characterised by deafness, vertigo and usually tinnitus which has its pathologic correlate hydropic distension of the endolymphatic system"[62]. The definition included the concept of endolymphatic hydrops as an integral part of Meniere's disease, otherwise the definition is unchanged from the description given by Prosper Meniere. The Academy also recognised correctly 2 major variants of Meniere's disease, vestibular Meniere's disease (vestibular hydrops) and cochlear Meniere's disease (cochlear hydrops). Two additional variants of Meniere's disease are also recognised - the Lermoyez syndrome and the drop attack or otolithic crisis (of Tumarkin) which are presumed to be secondary to endolymphatic hydrops[63]. Except for these few refinements, little has changed in the classic description of Meniere's disease in over 100 years. Meniere's disease is characterised by discontinuity and variability in the 3 major symptoms, viz vertigo, deafness and tinnitus.

The most spectacular feature is vertigo which may be accompanied by vomiting; the most disabling feature is the deafness which may leave the patient with a serious residual hearing defect if early treatment is not instituted, but the tinnitus may be the dominant feature among the symptoms. This may be the prodromal sympton heralding an attack and may almost constitute an "aura".

Attacks of Meniere's disease are intermittent and may follow one another with intervals of days or several months. As the deafness increases, the attacks gradually become less severe and may finally disappear completely after several years by which time the patient's deafness has become severe or total.

The true incidence of Meniere's disease is not known[78-80]. Neither is its prevalence. The actual spectrum of this inner ear disorder and its natural history is poorly understood except for an indeterminate number of clinically

moderate to severe cases. Estimates of the incidence of Meniere's disease have varied greatly because of the inconsistency in establishing the diagnosis by primary physicians. One of the greatest problems is that the initial presentation of the disease is often the cochlear form, which goes unrecognised. Even after the vestibular component becomes obvious, long periods of remission may mask the final full blown picture of episodic vertigo, fluctuant hearing loss, tinnitus and aural fullness. Thus only the clinically moderate to severe cases have been tabulated in estimates to date. Little is known of the extent of this disease process in its less severe forms.

Various estimations of the incidence have been reported. Harrison and Naftalin in 1968[8] suggested 1 per 1,000 population in Britain, whereas Cawthorne and Hewlett in 1954[64] made a much higher estimate of 1 per 636 population in Great Britain. Stahle et al in 1978[65] from a computer based study with a sample size of 28% of the total population, produced a much lower estimate of 46 per 100,000 population. This latter study is, however, probably on the conservative side because only patients who satisfied strict criteria were diagnosed as Meniere's disease, ie the more severe forms.

Many people with early Meniere's disease may never be evaluated by an otolaryngologist because the primary physician contact may not believe there is any effective treatment other than symptomatic therapy. At least 60% of afflicted persons may have significant clinical remissions of their symptoms, but if thoroughly tested are found to have progressive vestibular dysfunction[66].

The disease is equally common in the 2 sexes and the age of onset is also similar between males and females. It is unusual for idiopathic endolymphatic hydrops to develop after the age of 60 years[68,69].

In the literature the incidence of bilateral Meniere's disease has been estimated at between 2 to 78% of cases[70,74]. In the largest study to date 610 patients with Meniere's disease were followed up for 5 years. One hundred and eighty suffered bilateral involvement, more than half were afflicted within 5 years after the onset of initial unilateral symptoms. The onset of bilateral

symptoms in 103 patients came within a 2 year period and a further 23 patients were affected within 5 years; which in total makes 70% of patients affected by bilateral disease within 5 years. The average age of onset of disease in this group was between 50 and 54 years of age[71]. The time interval before bilateral involvement may differ significantly, eg in a series of Swedish studies after 3 years, 8% of patients with Meniere's disease had bilateral involvement. This figure rose to 25% after 10 years[72]. If vestibular function tests are done systematically, bilateral abnormalities are the rule rather than the exception, whether the cochlear loss is unilateral or bilateral[116]. Perhaps, therefore, greater efforts should be made at an earlier time to investigate the possibility of bilateral disease[73].

The variety of other diseases which may be manifest under the umbrella of Meniere's syndrome is legion. William House of Los Angeles wrote in 1975[67] that he makes the diagnosis of Meniere's disease less and less often. As interest and knowledge of the dizzy patient evolves, many specific causes for dizziness have been found. The term Meniere's syndrome, thus, is better used to include all conditions which include the triad of dizziness, with associated deafness and tinnitus, whereas Meniere's disease should be reserved for those patients who suffer from the condition known as endolymphatic hydrops.

House, therefore, requires a careful history from the patient before proceeding to laboratory tests. He requires the dizziness to be episodic with definite asymptomatic spells between attacks, in contrast to the dizzy patient who has a balance problem, with either good and bad days or constant dysequilibrium. The patient with far advanced hydrops may, however, be constantly slightly unsteady because of permanent inner ear damage. The vertigo should be associated with diminished hearing in the involved ear. The hearing should improve between attacks. In most cases the patient has fluctuating tinnitus and increased ear pressure with his attacks. Morrison, in contradiction of the above, believes 7% of Meniere's cases present with vertigo as the initial and early symptom and with a history of longer than 6 months[36,69].

The tinnitus in Meniere's disease is best characterised as a low-pitched, narrow band of noise usually described as a "roaring sound"[75,77]. The variability of the vertiginous attacks in Meniere's disease may have drastic effects on the life-style of the afflicted patient. Patients tend to become obsessed with the disease, which may cause agitation or depression to the point where the physician may feel there is a strong psychological component exaggerating the symptoms[81,106-108]. Indeed, some physicians believe the abnormal personality or psychiatric illness is part of the aetiology of the illness[114-116]. In a careful analysis of the literature, however, relating psychologic factors to Meniere's disease, vertigo or tinnitus, Crary and Wexler revealed a variety of methodologic flaws[82]. These authors later compared Meniere's patients with vertiginous control subjects without Meniere's disease in whom the aetiologies were known. In comparing these 2 groups by a wide variety of personality measures, no evidence was found of psychophysiologic processes in patients with Meniere's disease[83]. However, Pulec makes the flat statement that emotional or psychiatric factors are not involved in the aetiology[109].

The aetiology of Meniere's disease remains obscure. Although he was rumoured to be in the process of ascribing a cause for this disease, Meniere might be remembered for his perspicacity for not doing so.

Two main theories have been advanced to explain the presence of endolymphatic hydrops. One theory suggests a disturbance in the formation of the inner ear fluids which results in an increase in the endolymphatic compartment. The radial flow theory of endolymph movement suggests that the stria vascularis absorbs the endolymph which is derived from surrounding perilymph. Any decrease in the production of perilymph, therefore, results in an apparent increase of endolymph which later becomes a true increase as electrolyte content in the endolymph increases. Some authors feel the basic defect to be one of excess histamine being present in the stria vascularis, causing a vasodilation, increase in capillary permeability and hydrops[84]. Underlying labyrinthine ischaemia has also been thought for some time to be the basic defect present.

Seymour suggests that this results in local anoxia and production of a decreased amount of endolymph of high osmolarity[85]. Fluid is then transferred by osmosis from the surrounding perilymph and vascular spaces to the endolymph.

The other main theory is one which suggests the underlying problem is one of malfunction of the endolymphatic sac which may reabsorb the endolymph[86]. The microscopic appearance of the endolymphatic sac in animals is similar to that found in structures with an absorbative function[88] and it is thought that loss of this function results in an accumulation of endolymph and, therefore, hydrops[89]. Other proposals for the aetiology of the disease have included avitaminosis[90], viral infection[91] and endocrine disturbances[92] but none have gained widespread acceptance.

The possibility that allergic angioneurotic oedema might be one of the causes of endolymphatic hydrops was first considered by Quincke in 1893[96] but it was not until 1923 that Duke presented evidence for a specific allergic cause in Meniere's disease. Since that time further evidence has accumulated incriminating immediate hypersensitivity where arterial or spasm produces anoxia of the capillary loop and releases histamine which leads to dilatation and an increase in permeability[93,97]. The subsequent increase in endolymph protein could produce hydrops, particularly in predisposed labyrinths where a degree of water balance already exists[98]. Spasm of smooth muscle in branches of the internal auditory artery supplying the cristae and maculae, will produce local anoxia and exacerbate the vertigo.

Although atopy is thought to be common in these patients[99], it is the specific food allergies that have gained so much attention[96,100]. Clinically 2 types of food allergy were identified by Rinkel et al[101] - fixed food sensitivity and cyclic food sensitivity. In fixed food sensitivity, the same symptoms occur each time the food is eaten. The degree of sensitisation is not altered by ingestion or prolonged elimination of food. The problem is more complex in cyclic food allergy where the elimination of the food leads to tolerance or adaptation whilst frequent ingestion results in greater sensitisation.

Diagnosis is often difficult, however, especially from the history, since the symptoms of food allergy are often vague and include flatulence, fluid retention, post-prandial fatigue, epigastric discomfort, headaches and nausea[96]. Many food stuffs have been mentioned including dairy produce, wheat, corn, meat and vegetables, but there seems little agreement in the most common offending foods. Sensitivity to chocolate is an interesting example because it can also cause migraine[99] and the association between Meniere's disease and migraine has been well documented[74,102,103].

Many other observations relevant to aetiology have been made. Attention has been redirected to the possible relation of middle ear pressure abnormalities and the development of Meniere's disease. There is a higher incidence of negative middle ear pressures in Meniere's disease subjects than normals. The establishment of a middle ear pressure gradient of plus 60 cm water relevant to the ambient room pressure causes nystagmus and vertigo in normal subjects, but underpressure to the middle ear does not induce any vestibular reactions. During an acute attack the symptoms of vertigo, tinnitus, fullness and hearing loss are eliminated or reduced in 60% of patients by a reduction of the atmospheric pressure in a pressure chamber. However, if the negative middle ear pressure is a dominant aetiologic factor in Meniere's disease, the disorder should be common in children[3]. Tumarkin[110,111] began a cult of inserting a grommet through the tympanic membrane for the relief of vertigo in Meniere's disease, but Cinnamond[112] and Herman et al[113] totally refuted this negative middle ear pressure theory leading to Meniere's disease.

CONCLUSION

It is evident that knowledge of Meniere's disease is fragmentary but expanding. The aetiology of the disease remains obscure, the pathophysiology is uncertain and the clinical manifestation variable. In the midst of this instability no single line of treatment for the patient with Meniere's disease can be fully supported nor condemned until the foundations of knowledge are solidly laid.

REFERENCES

1 WILSON T D M, (1975) Otolaryngologic Clinics of North America 8 (2):259

2 MENIERE M P, (1861) Gaz Med Paris 16:55,88,239,279,597

3 HARKER L A and McCABE B F, (1980) Otolaryngology II 1978
W B Saunders Company, Philadelphia

4 ATKINSON M, (1961) Acta Otolaryngologica Supplement 162:1-78

5 KNAPP H, (1871) Archives Ophthalmol 2:204

6 DONALDSON J A, (1975) Otolaryngologic Clinics of North America 8 (2):267

7 HALLPIKE C S and CAIRNS H, (1938) Journal of Laryngology and Otology 53:625

8 HARRISON S M and NAFTALIN L, (1968) Meniere's Disease
Charles C Thomas Springfield Illinois

9 HALLPIKE C S and WRIGHT A J, (1940) Journal of Laryngology and Otology 55:59

10 ROLLIN H, (1940) Z HALS-NAS Ohrenheilk 3:72

11 ALTMANN F and FOWLER E P, (1943) Ann Otology 52:52

12 LINDSAY J R, (1953) Trans Amer Acad Ophthal Otolaryng 57:27

13 HALLPIKE C S, (1955) Post Graduate Medical Journal 31:330

14 FRIEDMANN I, (1974) Pathology of the Ear, Blackwell Scientific Publications
Page 476

15 ALTMAMM F and KORNFIELD M, (1965) Ann Otology 74:915

16 SCHUKNECHT H F, (1975) Otolaryngologic Clinics of North America 8 (2):507

17 KARMODY C S and SCHUKNECHT H F, (1966) Archives of Otolaryngology 83:18

18 KORNFIELD M, (1967) Pract Oto-Rhino-Laryng 29:406

19 RUEDI L, (1968) Acta Otolaryngologica 65:13

20 BRETLAU P and JORGENSEN M G, (1968) Journal of Laryngology and Otology 82:65

21 JOHNSSON L and HAWKINS J, (1973) Excerpta Medica 276:177

22 McCABE B F, (1966) Proc Trans Pacific Oto-Ophth Soc 47:37

23 BLACK F O, SANDO I, HILYARD V H and HEMMEN WAY W G, (1969) Ann Otology 78:1062

24 ALTMANN F and ZECHNER G, (1968) Arch Klin Exp Ohr-Nas Kehlk Heilk 192:1

25 SHAMBAUGH G E, (1969) Archives Otology 89:124

26 SHAMBAUGH G E and WIET R J, (1980) Otolaryngologic Clinics of North America
13 (4):585

27 VALVASSORI G E and CLEMIS J D, (1968) Otolaryngologic Clinics of North America 1:339

28 SACKETT J F, KOZAREK J A, ARENBERG I K, (1980) Otolaryngologic Clinics of North America 13:657

29 ARENBERG I K, GADO M H and SPECTOR G J, (1976) Rev Laryng 97:765

30 SHAMBAUGH G E, CLEMIS J D and ARENBERG I K, (1971) Archives Otolaryngology 89:38

31 LAWRENCE M and YANTIS P A, (1957) Ann Otology 66:595

32 PORTMANN G, (1927) Journal of Laryngology and Otology 42:809

33 PORTMANN G, (1980) Otolaryngologic Clinics of North America 13 (4):567

34 KIMURA R S and SCHUKNECHT H F, (1965) Pract Otorhinolaryng 27:343

35 KIMURA R S, (1967) Ann Otology 76:664

36 MORRISON A W, (1981) Proceedings of the Royal Society of Medicine 74:183

37 VON BEKESY G, (1952) Journal of the Acoustical Society of America 24:72

38 SMITH C A, LOWRY O H and WU H L, (1954) Laryngoscope 64:141

39 GUILD S R, (1927) American Journal of Anatomy 39:57

40 ORMOROD F C, (1960) Journal of Laryngology and Otology 74:659

41 LINDSAY J R, (1947) Archives of Otolaryngology 45:1

42 LINDSAY J R, SCHUKNECHT H F, NEFF W D and KIMURA R S, (1952) Ann Otology 61:697

43 SCHUKNECHT H F and KIMURA R S, (1953) Laryngoscope 63:1170

44 NAFTALIN L and HARRISON M S, (1958) Journal of Laryngology and Otology 72:118

45 LAWRENCE M, WOLSK D and LITTON W B, (1961) Ann Otology 70:753

46 LAWRENCE Mand CLAPPER M, (1961) Stain Technol 36:305

47 LAWRENCE M, (1964) Archives Otology 79:336

48 LAWRENCE M, (1960) Ann Otology 69:480

49 DAVIS N, THSAKI I, SMITH C A and DEATHERAGE B N, (1955) Fed Proc 14:35

50 LAWRENCE M, (1981) Otolaryngologic Clinics of North America 13:557

51 WULLSTEIN H L and RANCH S, (1961) Archives of Otolaryngology 73:262

52 TONNDORF J, (1957) Ann Otology 66:766

53 McCABE B F and WOLSK D, (1961) Ann Otology 70:541

54 HENRIKSSON N C, (1968) Otolaryngologic Clinics of North America 1:363

55 PIETRANTONI L and IURATO S (1960), Acta Otolaryngologica 52:15

56 LITTON W B and LAWRENCE M (1961), Archives of Otolaryngology 74:32

57 IRELAND P E and FARKASHIDY J (1963), Trans Amer Acad Ophthal Otolaryng 67:28

58 FRIEDMANN I, CAWTHORNE T and BIRD E S (1965), Journal Ultra Structure Res 12:92

59 FRIEDMANN I, CAWTHORNE T and BIRD E S (1965), Nature 207:171

60 HILDING D A and HOUSE W F (1964),Laryngoscope 74:1135

61 ALTMANN F (1961), Journal American Medical Association 176:215

62 ALFORD B R (1972) Trans Amer Acad Ophthalmol Otolaryngol 76:1462

63 SCHUKNECHT H F (1978), Journal Cont Ed O R L Allergy 40:18

64 CAWTHORNE T and HEWLETT A B (1954) Proceedings of Royal Society of Medicine 47:66

65 STAHLE J, STAHLE C and ARENBERG I K (1978), Archives of Otolaryngology 104:99

66 ARENBERG I K, BALKANY T J, GOLDMAN G and PILLSBURY H C (1980), Otolaryngologic Clinics of North America 13:597

67 HOUSE W F (1975), Otolaryngologic Clinics of North America 8:515

68 CAWTHORNE T (1947), Ann Otology 56:18

69 MORRISON A W (1975) Management of Sensori-neural Deafness, Butterworth & Co London

70 KITAHARA M, MATSUBARAH H, TAKEDA T and YAZAWA Y (1979), Adv Otorhinolaryngol 75:117

71 THOMAS K and HARRISON M S (1971) Proceedings of Royal Society of Medicine 66:853

72 STAHLE J and BERGMAN B (1967), Laryngoscope 9:1629

73 BALKANY T J, SIRES B and ARENBERG I K (1980) Otolaryngologic Clinics of North America 13:603

74 GOLDING-WOOD P H (1960), Journal of Laryngology and Otology 74:803

75 GRAHAM J T and NEWBY H A (1962), Archives of Otolaryngology 75:162

76 NODAL R H and GRAHAM J T (1965), Archives of Otolaryngology 82:28

77 REED G F (1960), Archives of Otolaryngology 71:94

78 EDWARDS C H (1971), Scott-Brown's Diseases of the Ear, Nose and Throat Third Edition, Butterworth & Co London

79 WILMOT T J (1973), Proceedings of Royal Society of Medicine Presidential Address

80 CAWTHORNE T (1965), Journal of Laryngology and Otology 70:695

81 LACHMAN S J (1972), Psychosomatic Disorders: A Behaviourial Investigation John Wiley & Sons Inc, New York

82 CRARY W G and WEXLER M (1977), Psychol Rep 1:V41

83 HOUSE J W, CRARY W G and WEXLER M (1980), Otolaryngologic Clinics of North America 13:625

84 WILLIAMS L (1965) Laryngoscope 75:1661

85 SEYMOUR J C (1960), Journal of Laryngology and Otology 74:559

86 BAST T H and ANSON B J (1950), Ann Otology 59:1088

87 BEAL D (1968), Acta Otolaryngologica 66:333

88 LINDQUIST P G, KIMURA R and WERSALL J (1964), Acta Otolaryngologica 57:65

89 ARENBERG I,MAROVITZ W and SHAMBAUGH G (1970), Acta Otolaryngologica Suppl 275

90 SELFRIDGE G (1940), Ann Otology 72:687

91 LEMPERT J, WOLFF D, RAMBO J,WEVER E and LAWRENCE M (1952), Ann Otology 61:717

92 GOLDMAN H B (1962), New York Journal of Medicine 63:377

93 SHAMBAUGH G E and WIET R J (1980), Otolaryngologic Clinics of North America 13:671

94 RAUCH S and KOSTALIN A (1962), 2 Laryng Rhin Otol 41:56

95 DUVALL A J and RHODES U T (1967), Ann Otology 76:688

96 SHAVER E F (1975), Archives of Otolaryngology 101:96

97 WILLIAM M L (1952), Meniere's Disease Edition One, Charles C Thomas Springfield, Illinois

98 MYGIND S H and DEDERDING D (1938), Ann Otology 47:56

99 CRIEP L H (1939), Pennsylvania Medical Journal 43:258

100 POWERS W H and HOUSE W F (1969), Laryngoscope 79:1330

101 RINKEL H J, RANDOLPH T J and ZELLAR M (1951), Food Allergy, Charles C Thomas Springfield, Illinois

102 ATKINSON M (1943), Annals Internal Medicine 18:797

103 HINCHCLIFFE R (1967), Acta Otolaryngologica 63:384

104 GODLOWSKI Z (1972), Acta Otolaryngologica Suppl 299

105 WILMOT T J (1974), Journal of Royal Society of Medicine 67:331

106 HINCHCLIFFE R (1967), Journal of Laryngology and Otology 81:471

107 HINCHCLIFFE R (1967), Journal of Laryngology and Otology 81:477

108 HINCHCLIFFE R (1967), Journal of Laryngology and Otology 81:661

109 PULEC J L (1972) Laryngoscope 82:1703

110 TUMARKIN A (1961), Journal of Royal Society of Medicine 54:907

111 TUMARKIN A (1966), Journal of Laryngology and Otology 80:1041

112 CINNAMOND M J (1975), Journal of Laryngology and Otology 89:57

113 HERMAN L E, STROUD M H, CARVER W F and ARENBORG K (1977), Archives of Otolaryngology 103:700

114 FOWLER P and ZECKEL A (1953), Psychosomatic Medicine 15:127

115 STEPHENS S D G (1975), Journal of Laryngology and Otology 89:479

116 WILMOT T J (1974), Journal of Laryngology and Otology 88:295